CASE REPORTS

PATENT DUCTUS ARTERIOSUS WITH VENTRICULAR SEPTAL DEFECT

BY

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The case described below is of interest in that the patient survived until the age of 69 with a double congenital lesion, complicated by essential hypertension.

Case Report

A woman, aged 58, was admitted to Snowdon Road Hospital, Bristol, in 1945, with a twenty-year history of dyspnœa, palpitation, occasional ankle swelling, and various neurotic symptoms. On examination, she was found to have kyphoscoliosis and emphysema. The pulse was regular; the apex beat was in the left sixth intercostal space beyond the midclavicular line. The heart sounds were normal, with a basal and central systolic murmur and thrill, and the blood pressure was 190/100 mm. Hg. The electrocardiogram then and again ten years later was normal. Chest X-ray showed cardiac enlargement of unspecified degree. These findings were noted by three consultant physicians who had examined her on various occasions about this period.

She remained, with the exception of a few months, in this hospital for the chronic sick until her death in 1956. Re-examined in 1955, she was found to be apprehensive, with symptoms of dyspnœa, orthopnœa, paroxysmal nocturnal dyspnœa, and dependent œdema. There was no cyanosis or finger clubbing. The venous pressure was raised, the jugular veins being distended to 2·5 cm. above the clavicles in the sitting position. The pulse rate was 100 a minute, regular, and collapsing in character. The blood pressure had risen to 260/140 mm. The apex beat was in the left sixth intercostal space at the anterior axillary line, heaving in character. The heart sounds were tic-tac; the pulmonary second sound was split. There was a loud machinery murmur in the pulmonary area, and also a separate systolic murmur with a thrill at the sternal border in the left fourth intercostal space.

The clinical diagnosis was patent ductus arteriosus with a ventricular septal defect, complicated by systemic hypertension and congestive heart failure. Her condition gradually deteriorated, and she died suddenly at the age of 69, in 1956, nearly 11 years after her admission.

Necropsy. The heart weighed 381 g. There was left ventricular hypertrophy (average thickness 12·5 mm.); the right ventricle was also hypertrophied (thickness 6 mm.); both chambers were dilated. The mitral, tricuspid, and pulmonary valves were normal; there was some adherence of the aortic cusps. Below the pulmonary valve, in the muscular part of the septum, near the fibromuscular junction, there was a defect, 18 mm. in diameter, in its right ventricular face, passing obliquely through the septum, so that it was 4 mm. at its left ventricular face. The aorta was not dilated, the circumference immediately above the valve being 67 mm. (normal=50–90 mm.); in addition there were severe atheromatous changes throughout its length, these being greatest in the abdominal aorta. The pulmonary artery, immediately above the pulmonary valve, was 48 mm. in circumference, with many intimal atheromatous patches; the smaller pulmonary arteries and arterioles showed no evidence of pulmonary hypertension. There was a patent ductus arteriosus, 17 mm. in length, running obliquely through adhesions between the aorta and left pulmonary artery. The aortic orifice, situated in a small localized dilatation, was 8×5 mm. in size. Around the pulmonary orifice, which was 2 mm. in diameter, was a spout-like elevation of the intima, about 1 mm. high. The kidneys showed changes of essential hypertension. The lungs showed bronchopneumonic changes.

Discussion

The two main features of interest in this patient are the age to which she survived and the apparent change in physical signs. The life span in patients with patent ductus arteriosus may be

normal and the following examples have been recorded, together with data of the ductus size. The size of the ductus and the size of the pressure gradient between the aorta and the pulmonary artery are the two factors that determine the size of the shunt, and hence the prognosis of the patient. It would seem that the smaller the ductus the better the prognosis as to life span.

Author	Patient's sex and age at death	Aperture of ductus	
		Aortic	Pulmonary
Josefson (1897)	F. 66	6×10 mm.	4 mm.
White (1928)	F. 65	10 mm.	4 mm.
Brody and Randall (1935)	F. 66	9 mm.	4 mm.
Walker and Ellis (1940)	M. 73	—	—
Fishman and Silverthorne (1951)	F. 75	3 mm.	4 mm.
Higgins (1958)	F. 69	8×5 mm.	2 mm.

In addition, in this patient there were two additional factors in that she had hypertension and a ventricular septal defect—factors that might reasonably be presumed to have shortened the life span. It is fair to point out that the patient had been sheltered in hospital for at least ten years, and that for the greater part of her life she had been an invalid, not so much as a result of her cardiac condition, but as a result of a chronic neurosis with feelings of insecurity dating from childhood.

The apparent change, if we accept it, of a basal systolic murmur to a machinery murmur also requires comment. The presence of right ventircular hypertrophy and pulmonary artery atheroma

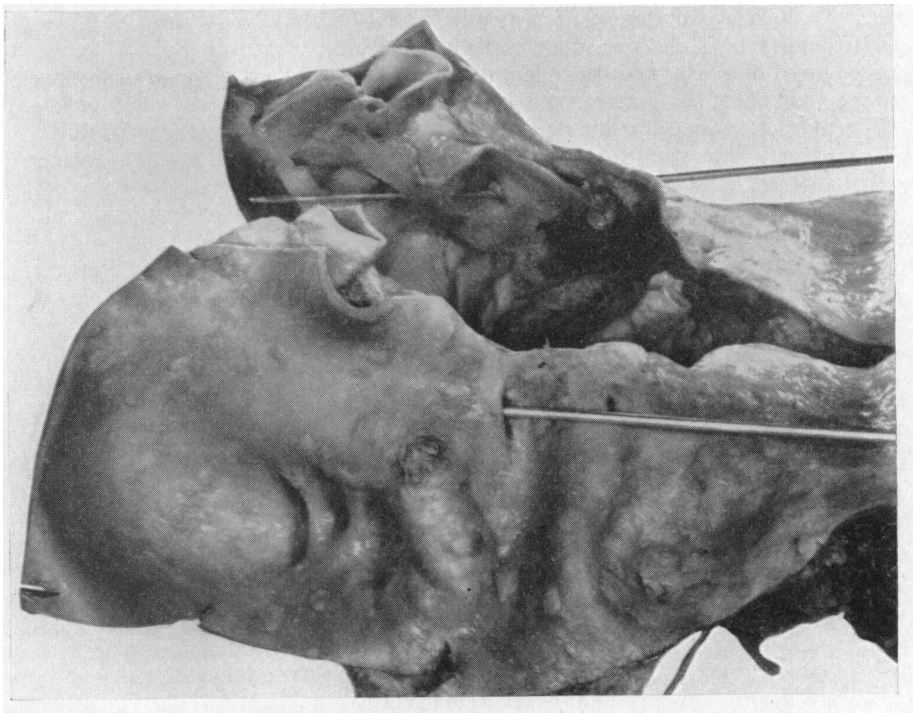

FIG. 1.—The aorta is seen in the foreground with the probe entering the aortic orifice of the ductus. Below and to the left are the openings of the innominate, left common carotid, and left subclavian arteries. The back of the specimen has been shown by arranging a mirror so that it is reflected, somewhat above the front view, and the same probe is seen leaving the pulmonary orifice of the ductus.

may be due not only to the shunts but also to pulmonary hypertension, although there were no changes in the pulmonary arterioles suggestive of this. If so, the pressure gradient between the aorta and pulmonary artery would be relatively low, and hence there might be a systolic murmur only. Later, when the systolic blood pressure was continuously in the region of 260/140 mm., the gradient became greater and led to the production of a continuous murmur. Alternatively, it seems possible that the continuous murmur was present but was not noted.

Summary

A woman with a persistent ductus arteriosus and a ventricular septal defect, who survived until the age of 69 although she had also essential hypertension, is described, together with autopsy findings.

Comments are made on the life span and the size of the ductus.

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